

# General

### Guideline Title

EFNS-ENS guidelines on the diagnosis and management of disorders associated with dementia.

### Bibliographic Source(s)

Sorbi S, Hort J, Erkinjuntti T, Fladby T, Gainotti G, Gurvit H, Nacmias B, Pasquier F, Popescu BO, Rektorova I, Religa D, Rusina R, Rossor M, Schmidt R, Stefanova E, Warren JD, Scheltens P, EFNS Scientist Panel on Dementia and Cognitive Neurology. EFNS-ENS guidelines on the diagnosis and management of disorders associated with dementia. Eur J Neurol. 2012 Sep;19(9):1159-79. [189 references] PubMed

### Guideline Status

This is the current release of the guideline.

This guideline updates a previous version: Waldemar G, Dubois B, Emre M, Georges J, McKeith IG, Rossor M, Scheltens P, Tariska P, Winblad B, EFNS. Recommendations for the diagnosis and management of Alzheimer's disease and other disorders associated with dementia: EFNS guideline. Eur J Neurol 2007 Jan;14(1):e1-26.

# Regulatory Alert

# FDA Warning/Regulatory Alert

Note from the National Guideline Clearinghouse: This guideline references a drug(s) for which important revised regulatory and/or warning information has been released.

May 10, 2016 – Olanzapine : The U.S. Food and Drug Administration (FDA) is warning that the antipsychotic medicine olanzapine can cause a rare but serious skin reaction that can progress to affect other parts of the body. FDA is adding a new warning to the drug labels for all olanzapine-containing products that describes this severe condition known as Drug Reaction with Eosinophilia and Systemic Symptoms (DRESS).

# Recommendations

# Major Recommendations

The levels of evidence (class I-IV) supporting the recommendations and levels of recommendation (A-C, Good Practice Point) are defined at the end of the "Major Recommendations" field.

### Diagnostic Evaluation

Clinical Diagnosis: Medical History, Laboratory, Neurological, and Physical Examination

Clinical history should be supplemented by an informant (Good Practice Point) (Hort et al., 2010; American Psychiatric Association (APA), Task Force on DSM-IV, 2000; International Statistical Classification of Diseases and Related Health Problems 10th Revision Version [ISCD] for 2007; Diagnostic Statistical Manual DSM 5 [DSM5], 2012). A neurological and general physical examination should be performed in all patients with dementia (Good Practice Point) (Hort et al., 2010; APA Task Force on DSM-IV, 2000; ISCD, 2007; DSM5, 2012; Rossor et al., 2010). Routine blood tests are useful in excluding co-morbidities (Good Practice Point) (Knopman et al., 2001).

Assessment of Cognitive Functions, Screening Tests, and Assessment of Specific Cognitive Domains

Cognitive assessment is central to diagnosis and management of dementias and should be performed in all patients (Level A) (Knopman et al., 2001). Screening tests are available of good accuracy in the general diagnosis of dementia or have been proposed specifically for the differential diagnosis between the different forms of dementia (Good Practice Point) (Knopman et al., 2001). Neuropsychological assessment should be performed in all patients in the early stages of the disease (Level B) when the cognitive impairment reflects the disruption of specific brain structures (Hort et al., 2010; APA Task Force on DSM-IV, 2000; ISCD, 2007; DSM5, 2012; Braak & Braak, 1991). The neuropsychological assessment should include a global cognitive measure, and in addition, more detailed testing of the main cognitive domains including memory, executive functions, and instrumental functions (Level C) (Knopman et al., 2001).

Assessment of Behavioural and Psychological Symptoms of Dementia (BPSD)

Assessment of BPSD is essential for both diagnosis and management and should be performed in each patient (Good Practice Point) (Aalten et al., 2008). Information is gathered from an informant using an appropriate rating scale (Good Practice Point) (Conn & Thorpe, 2007). Although specific BPSD form the core or supportive features of some non-Alzheimer's disease dementias, co-morbidity should always be considered as a possible cause (Good Practice Point) (Gorno-Tempini et al., 2011; Rascovsky et al., 2011; McKeith, 2006; Emre et al., 2007).

Assessment of Activities of Daily Living (ADL)

ADL and Instrumental ADL (IADL) impairment because of cognitive decline is an essential part of the diagnostic criteria for dementia and should be assessed in the diagnostic evaluation (Good Practice Point) (Galasko et al., 1997; Pfeffer et al., 1982; DeJong, Osterlund, & Roy, 1989; Lawton & Brody, 1969; Gelinas et al., 1999; Gleichgerricht et al., 2009; Mioshi & Hodges, 2009). A semi-structured interview from the caregiver is the most practical way to obtain relevant information, and various validated scales translated into different languages are available (Good Practice Point) (Galasko et al., 1997; Pfeffer et al., 1982; DeJong, Osterlund, & Roy, 1989; Lawton & Brody, 1969; Gelinas et al., 1999; Gleichgerricht et al., 2009; Mioshi & Hodges, 2009).

### Assessment of Co-morbidity

Assessment of co-morbidity is important in demented patients, both at the time of diagnosis and throughout the course of the illness (Good Practice Point) (Fu et al., 2004) and should always be considered as a possible cause of BPSD (Good Practice Point) (Meier & Lemcke, 2010). Blood levels of folate, vitamin B12, thyroid-stimulating hormone, calcium, glucose, complete blood cell count, renal and liver function tests should be evaluated at the time of diagnosis and serological tests for syphilis, Borrelia, and human immunodeficiency virus (HIV) might also be needed in cases with atypical presentation or clinical features suggestive of these disorders (Good Practice Point) (Knopman et al., 2001).

### Neuroimaging

### Structural Imaging

Structural imaging should be used in the evaluation of every patient affected by dementia (Level A) (Clarfield, 2003). Computed tomography (CT) and standard magnetic resonance imaging (MRI) are used to exclude secondary causes for dementia such as tumour, inflammatory disease, including abscess or normal-pressure hydrocephalus (Level A) (Clarfield, 2003). It is particularly difficult to attribute clinical significance to the evidence of cerebrovascular disease in patients with cognitive impairment. At the current state of knowledge, demonstration of cerebrovascular disease on imaging is used to support the diagnosis (Good Practice Point) (Van Straaten et al., 2003; Gold et al., 2002). Atrophy distribution is useful in the differential diagnosis of frontotemporal lobar degeneration (FTLD) compared with Alzheimer's disease (AD) and of the subtypes of FTLD (Level C) (Kipps et al., 2009; Galton et al., 2001; Kipps et al., 2007). No established structural MRI pattern is characteristic for dementia with Lewy bodies (DLB) and Parkinson's disease dementia (PDD) (Good Practice Point) (Burton et al., 2004). MRI is used to distinguish progressive supranuclear palsy (PSP) from DLB, being midbrain and the superior cerebellar peduncles atrophic in PSP (Good Practice Point) (Burton et al., 2004). The pronounced atrophy of the caudate nucleus and putamen is characteristic, and the so-called bicaudate ratio doubles of Huntington's disease (HD) (Level B) (Kuehn, 2011; Quattrone et al., 2008). MRI showing diffusion-weighted imaging (DWI) cortical rims, striatal

and/or thalamic hyperintensities is useful for the diagnosis of sporadic Creutzfeldt-Jakob disease (CJD) (Level A) (Zerr et al., 2009; Young et al., 2005). The MRI pulvinar sign, that is, symmetrical fluid attenuated inversion recovery (FLAIR) hyperintensity of the posterior thalamus, has high diagnostic utility for *variant* CJD (Level B) (Zeidler et al., 2000). Diffusion-tensor imaging (DTI) MRI distinguishes FTLD from AD and controls (and AD from controls) (Level B) (Avants et al., 2010; Whitwell et al., 2010). Measuring flow void on MRI can increase confidence on neuropsychiatric inventory (NPI) diagnosis and on decision about shunt placement (Good Practice Point) (Palm et al., 2006). Hyperintense signal abnormality on T2-weighted images within medial temporal lobe structures such as the hippocampi and amygdalae and, on occasion, the hypothalamus are commonly seen in limbic encephalitis (Level C).

### Functional Imaging Modalities

DTI MRI distinguishes frontotemporal dementia (FTD) from AD and controls (and AD from controls) (Level B) (Avants et al., 2010; Whitwell et al., 2010). DTI MRI shows the distinct patterns of diffusivity changes, in parkinsonistic disorders (PDD, DLB, progressive supranuclear palsy [PSP], corticobasal syndrome [CBS]) (Level C) (Whitwell et al., 2010). Single-photon emission computed tomography (SPECT) perfusion and MRI morphometric imaging are useful to distinguish DLB, CBS, CJD from AD (Good Practice Point) (Kantarci et al., 2010; Erbetta et al., 2009; Ukisu et al., 2005). SPECT pre-synaptic dopamine transporter imaging is useful to distinguish DLB from non-DLB dementias (Level B) (Goto et al., 2010; O'Brien et al., 2009). SPECT and positron emission tomography (PET) perfusion and metabolic techniques are highly useful in FTLD diagnosis (Mendez et al., 2007; Rabinovici et al., 2008; Josephs et al., 2010) (Level C).

### Electroencephalography (EEG)

EEG is recommended in *rapid dementia* and differential diagnosis when CJD or transient epileptic amnesia is suspected (Level B) (Jelic & Kowalski, 2009; Liedorp et al., 2009; Wieser, Schindler, & Zumsteg, 2006). There is not enough evidence to consider resting EEG for the initial assessment of all dementia patients.

### Cerebrospinal Fluid (CSF) Analysis

Routine CSF analysis may help to rule out or rule in certain infectious causes (Good Practice Point) (Jesse et al., 2011). CSF abeta 1-42/tau/p-tau assessment helps to differentiate AD (Level B) (Spies et al., 2010). Assessment of CSF total tau and 14-3-3 protein is recommended in rapidly progressive dementia when sporadic CJD (sCJD) is suspected (Good Practice Point) (Van Harten et al., 2011; Sanchez-Juan et al., 2006).

### Genetic Testing

No studies have addressed the value of genetic counselling for patients with dementia or their families when autosomal-dominant disease is suspected. Because the genetics of dementing illnesses is a very young field, expertise in genetic counselling for the dementias of the elderly is likely to be found only in specialized dementia research centres (Good Practice Point) (Goldman et al., 2011; Prusiner & Hsiao, 1994; "Guidelines for the molecular genetics," 1994). Screening for known pathogenic mutations can be undertaken in patients with appropriate phenotype or a family history of an autosomal-dominant dementia. This should only be undertaken in specialist centres with appropriate counselling of the patient and family caregivers, and with consent (Good Practice Point). Pre-symptomatic testing may be performed in adults where there is a clear family history, and when there is a known mutation in an affected individual to ensure that a negative result is clinically significant. It is recommended that the Huntington's disease protocol is followed (Good Practice Point).

### Biopsy and Other Investigations

Brain and other specific tissue biopsies can provide a diagnosis in rare or rapidly progressing dementias, but should only be carried out in specialist centres in carefully selected cases (Good Practice Point) (Schott et al., 2010; Warren et al., 2005).

### Management of the Dementias

### Primary and Secondary Prevention

No treatments, nor lifestyle, have demonstrated the efficacy for preventing or delaying the development of the different types of dementias until now.

### Treatment of Cognitive Deficits in Non-Alzheimer Dementias

Use of cholinesterase inhibitors (ChEIs), memantine or selective serotonin reuptake inhibitors (SSRIs) in any of the FTLD subtypes is possibly ineffective for cognitive improvement (Level C) (Bei et al., 2010; Lebert et al., 2004). Dopaminergic replacement with bromocriptine in progressive aphasias is probably ineffective (Good Practice Point) (Reed et al., 2004). Given the insufficient classes II and III evidence and the evidence being largely based on class IV, the use of ChEIs and memantine in FTLD cannot be recommended. There is little class III evidence in support of rivastigmine and memantine (Bei et al., 2010; Lebert et al., 2004). There is no independent evidence for recommending any therapeutic

intervention for CBS (Litvan et al., 2001; Zerr, 2009). Rivastigmine is the approved ChEI for the treatment of PDD with class I evidence. PDD diagnosis warrants the use of rivastigmine (Good Practice Point) (Maidment, Fox, & Boustani, 2006). Parallels with PDD in terms of clinical picture and disease mechanisms suggest that rivastigmine is possibly effective in DLB (GPP). The evidence for the efficacy of galantamine is insufficient for both PDD and DLB. Memantine is probably effective for both PDD and DLB (Level B) as there were consistently significant improvements in global measures, but not in cognitive measures in two class II studies (Aarsland et al., 2009; Emre et al., 2010). There is insufficient evidence for recommending any specific agent in the treatment of human prion diseases. Surgical treatment can be considered in normal pressure hydrocephalus (NPH) (Level C), and risk to benefit ratio must be individualized for each patient (Marmarou et al., 2005; Esmonde & Cooke, 2002). There is insufficient evidence for recommending any of non-pharmacological treatments.

### Treatments of BPSD

Antipsychotic medications, conventional and atypical agents, may be utilized in clinical practice for aggression, psychosis, and agitation as well as SSRIs for mood and behavioural disorders (Good Practice Point) (Ballard & Corbett, 2010); however, there is little evidence to guide practice.

### Counselling and Support for Caregivers

A dementia diagnosis mandates an inquiry to the community for available public health care support programs (Good Practice Point) (Ballard & Corbett, 2010). Counselling and case/care management amongst caring family members have positive effects on burden and satisfaction for caregivers of people with dementia (Good Practice Point).

### Decision-Making and Participating in Research

Research involving persons affected by dementia needs to adopt special precautions, and there is consensus over the fact that adults who lack capacity should be supported by proxy consent when involved in research (Good Practice Point) (Gainotti et al., 2010).

### Driving

Assessment of driving ability should be made after diagnosis with particular attention paid to visuo-spatial, visuo-perceptual, and executive abilities (Good Practice Point). Advice either to allow driving, but to review after an interval, to cease driving, or to refer for retesting should be given (Good Practice Point) (Adler & Rottunda, 2011).

### **Definitions**:

Evidence Classification Scheme for a Diagnostic Measure

Class I: A prospective study in a broad spectrum of persons with the suspected condition, using a 'gold standard' for case definition, where the test is applied in a blinded evaluation, and enabling the assessment of appropriate tests of diagnostic accuracy

Class II: A prospective study of a narrow spectrum of persons with the suspected condition, or a well-designed retrospective study of a broad spectrum of persons with an established condition (by 'gold standard') compared to a broad spectrum of controls, where test is applied in a blinded evaluation, and enabling the assessment of appropriate tests of diagnostic accuracy

Class III: Evidence provided by a retrospective study where either persons with the established condition or controls are of a narrow spectrum, and where test is applied in a blinded evaluation

Class IV: Any design where test is not applied in blinded evaluation OR evidence provided by expert opinion alone or in descriptive case series (without controls)

Evidence Classification Scheme for a Therapeutic Intervention

Class I: An adequately powered prospective, randomized, controlled clinical trial with masked outcome assessment in a representative population or an adequately powered systematic review of prospective randomized controlled clinical trials with masked outcome assessment in representative populations. The following are required:

- a. Randomization concealment
- b. Primary outcome(s) is/are clearly defined
- c. Exclusion/inclusion criteria are clearly defined
- d. Adequate accounting for dropouts and crossovers with numbers sufficiently low to have minimal potential for bias
- e. Relevant baseline characteristics are presented and substantially equivalent among treatment groups or there is appropriate statistical adjustment for differences

Class II: Prospective matched-group cohort study in a representative population with masked outcome assessment that meets a—e above or a randomized, controlled trial in a representative population that lacks one criteria a—e

Class III: All other controlled trials (including well-defined natural history controls or patients serving as own controls) in a representative population, where outcome assessment is independent of patient treatment

Class IV: Evidence from uncontrolled studies, case series, case reports, or expert opinion

Rating of Recommendations for a Diagnostic Measure

Level A rating (established as useful/predictive or not useful/predictive) requires at least one convincing class I study or at least two consistent, convincing class II studies.

Level B rating (established as probably useful/predictive or not useful/predictive) requires at least one convincing class II study or overwhelming class III evidence.

Level C rating (established as possibly useful/predictive or not useful/predictive) requires at least two convincing class III studies.

Good Practice Points (GPP) Where there was a lack of evidence, but clear consensus, good practice points are provided.

Rating of Recommendations for a Therapeutic Intervention

Level A rating (established as effective, ineffective, or harmful) requires at least one convincing class I study or at least two consistent, convincing class II studies.

Level B rating (probably effective, ineffective, or harmful) requires at least one convincing class II study or overwhelming class III evidence.

Level C rating (possibly effective, ineffective, or harmful) requires at least two convincing class III studies.

Good Practice Points (GPP) Where there was a lack of evidence, but clear consensus, good practice points are provided.

# Clinical Algorithm(s)

None provided

# Scope

# Disease/Condition(s)

Dementia disorders

Note: This guideline does not address Alzheimer's disease. See the EFNS guidelines for the diagnosis and management of Alzheimer's disease.

# Guideline Category

Counseling

Diagnosis

Evaluation

Management

Screening

Treatment

# Family Practice Geriatrics Internal Medicine Medical Genetics

Clinical Specialty

Neurology

Psychiatry

Radiology

### **Intended Users**

Physicians

# Guideline Objective(s)

- To present a peer-reviewed evidence-based statement for the guidance of practice for clinical neurologists, geriatricians, psychiatrists, and other specialist physicians responsible for the care of patients with dementing disorders
- To present a statement of minimum desirable standards for practice guidance

### **Target Population**

Patients with suspected or diagnosed dementia other than Alzheimer's disease

### Interventions and Practices Considered

### Evaluation/Diagnosis

- 1. Medical history
- 2. Neurological and physical examination
- 3. Blood tests
- 4. Assessment of cognitive function
- 5. Screening tests
- 6. Assessment of behavioral and psychological symptoms
- 7. Assessment of activities of daily living
- 8. Assessment of co-morbidities
- 9. Structural and functional neuroimaging (e.g., computed tomography [CT], magnetic resonance imaging [MRI], single photon emission computed tomography [SPECT], and positron emission tomography [PET])
- 10. Electroencephalography
- 11. Cerebrospinal fluid analysis
- 12. Genetic testing
- 13. Tissue biopsy in carefully selected cases
- 14. Assessment of driving ability

### Management/Treatment

- 1. Cholinesterase inhibitors (ChEIs) (e.g., rivastigmine)
- 2. Memantine (alone or in combination with ChEIs)
- 3. Monitoring treatment with ChEIs and memantine

- 4. Bromocriptine in progressive aphasias (probably ineffective)
- 5. Surgical treatment for normal-pressure hydrocephalus (NPH)
- 6. Selective serotonin reuptake inhibitors (SSRIs)
- 7. Conventional and atypical antipsychotics
- 8. Counseling and support for caregivers

Note: The following treatments were considered but not recommended due to insufficient evidence: primary or secondary prevention of dementia, any treatment for corticobasal syndrome (CBS) or prion disease, galantamine for Parkinson's disease dementia (PDD) and dementia with Lewy bodies (DLB), any non-pharmacological treatments for non-Alzheimer's dementia.

# Major Outcomes Considered

- Changes in the symptoms associated with non-Alzheimer dementias, including cognitive function, behavioral symptoms, functionality, and quality of life
- Accuracy of screening tests
- · Sensitivity and specificity diagnostic tests
- Efficacy of treatment interventions (measured by e.g., improvement in symptoms, scores on assessment tests, level of comorbidities)
- Incidence and severity of adverse effects of treatments
- Level of burden and satisfaction of caregivers

# Methodology

### Methods Used to Collect/Select the Evidence

Searches of Electronic Databases

# Description of Methods Used to Collect/Select the Evidence

The evidence for these guidelines was collected from Cochrane Library reviews, other published meta-analyses and systematic reviews, evidence-based management guidelines in dementia, and original scientific papers published in peer-reviewed journals before June 2011. The search strategy sought only studies published in English. The principal search term was dementia. Other terms entered into the search included diagnosis, guideline, management, recommendation, review, treatment. For each topic, the evidence was sought in MEDLINE according to predefined search protocols.\* Final inclusion of articles in this practice parameter was based on consensus of the committee.

\*Searching terms used in the search strategy: vascular cognitive impairment, frontotemporal lobar degeneration, dementia with Lewy bodies, corticobasal syndrome, progressive supranuclear palsy, Parkinson's disease dementia, Huntington disease, prion diseases, normal pressure hydrocephalus, limbic encephalitis, dementia, CSF, MR, Spect, FDG-PET, amyloid-PET, genetics, biopsy, DNA, EEG, dementia, and ethics.

### Number of Source Documents

Not stated

# Methods Used to Assess the Quality and Strength of the Evidence

Weighting According to a Rating Scheme (Scheme Given)

# Rating Scheme for the Strength of the Evidence

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Class II: A prospective study of a narrow spectrum of persons with the suspected condition, or a well-designed retrospective study of a broad spectrum of persons with an established condition (by 'gold standard') compared to a broad spectrum of controls, where test is applied in a blinded evaluation, and enabling the assessment of appropriate tests of diagnostic accuracy

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Evidence Classification Scheme for a Therapeutic Intervention

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Class II: Prospective matched-group cohort study in a representative population with masked outcome assessment that meets a—e above or a randomized, controlled trial in a representative population that lacks one criteria a—e

Class III: All other controlled trials (including well-defined natural history controls or patients serving as own controls) in a representative population, where outcome assessment is independent of patient treatment

Class IV: Evidence from uncontrolled studies, case series, case reports, or expert opinion

# Methods Used to Analyze the Evidence

Review of Published Meta-Analyses

Systematic Review

# Description of the Methods Used to Analyze the Evidence

The main goal of the task force was to determine whether further evidence had become available relating to biomarkers such as magnetic resonance imaging (MRI), positron emission tomography (PET) and cerebrospinal fluid (CSF) and to determine the evidence for use in practice. Special attention was given to the results of recent clinical trials, both for cognitive and behavioural aspects of the disease.

The scientific evidence for diagnostic investigations and treatments was evaluated according to pre-specified levels of certainty (class I, II, III and IV) (see the 'Rating Scheme for the Strength of the Evidence' field).

### Methods Used to Formulate the Recommendations

Expert Consensus

# Description of Methods Used to Formulate the Recommendations

The recommendations were graded according to the strength of evidence (grade A, B, or C), using the definitions given in the European Federation of Neurological Societies (EFNS) guidance. In addressing the important clinical questions, for which no evidence was available, the task force group recommended 'good practice points' based on the experience and consensus of the task force group.

A proposed guideline with specific recommendation was drafted for circulation to task force members and displayed on EFNS web pages for comments from all panel members. Consensus was reached at three task force meetings during 2010 and 2011 and through five revisions via the web.

# Rating Scheme for the Strength of the Recommendations

Rating of Recommendations for a Diagnostic Measure

Level A rating (established as useful/predictive or not useful/predictive) requires at least one convincing class I study or at least two consistent, convincing class II studies.

Level B rating (established as probably useful/predictive or not useful/predictive) requires at least one convincing class II study or overwhelming class III evidence.

Level C rating (established as possibly useful/predictive or not useful/predictive) requires at least two convincing class III studies.

Good Practice Points (GPP) Where there was a lack of evidence, but clear consensus, good practice points are provided.

Rating of Recommendations for a Therapeutic Intervention

Level A rating (established as effective, ineffective, or harmful) requires at least one convincing class I study or at least two consistent, convincing class II studies.

Level B rating (probably effective, ineffective, or harmful) requires at least one convincing class II study or overwhelming class III evidence.

Level C rating (possibly effective, ineffective, or harmful) requires at least two convincing class III studies.

Good Practice Points (GPP) Where there was a lack of evidence, but clear consensus, good practice points are provided.

# Cost Analysis

A formal cost analysis was not performed and published cost analyses were not reviewed.

### Method of Guideline Validation

Peer Review

# Description of Method of Guideline Validation

The guidelines were validated according to the European Federation of Neurological Societies (EFNS) criteria (see the "Availability of Companion Documents" field).

# Evidence Supporting the Recommendations

# References Supporting the Recommendations

Aalten P, Verhey FR, Boziki M, Brugnolo A, Bullock R, Byrne EJ, Camus V, Caputo M, Collins D, De Deyn PP, Elina K, Frisoni G, Holmes C, Hurt C, Marriott A, Mecocci P, Nobili F, Ousset PJ, Reynish E, Salmon E, Tsolaki M, Vellas B, Robert PH. Consistency of neuropsychiatric syndromes across dementias: results from the European Alzheimer Disease Consortium. Part II. Dement Geriatr Cogn Disord. 2008;25(1):1-8. [32 references] PubMed

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# Type of Evidence Supporting the Recommendations

The type of supporting evidence is identified and graded for selected recommendations (see the "Major Recommendations" field).

# Benefits/Harms of Implementing the Guideline Recommendations

### Potential Benefits

Appropriate diagnosis and management of disorders associated with dementia other than Alzheimer's disease

### **Potential Harms**

- Whilst patients with dementia associated with Lewy bodies respond to cholinesterase inhibitors with improvement in cognitive and
  psychiatric symptoms, they show a propensity to have exaggerated adverse reactions to neuroleptic drugs, with a significantly increased
  morbidity and mortality.
- Concerns about rivastigmine tolerability were stated in a Cochrane Library review on cholinesterase inhibitors treatment in Parkinson's disease dementia.
- Benefits from antipsychotic medications, conventional and atypical agents, for behavioural and psychological symptoms of dementia have to be considered in the context of significant adverse events, including extrapyramidal symptoms, accelerated cognitive decline, stroke, and death
- Surgical treatment for normal-pressure hydrocephalus carries considerable short- and long-term risks.

# Qualifying Statements

# **Qualifying Statements**

- This guideline provides the view of an expert task force appointed by the Scientific Committee of the European Federation of Neurological Societies (EFNS). It represents a peer-reviewed statement of minimum desirable standards for the guidance of practice based on the best available evidence. It is not intended to have legally binding implications in individual cases.
- These guidelines represent desirable standards, but may not be appropriate in all circumstances as clinical presentation of the individual patient and available resources should be taken into account.

# Implementation of the Guideline

# Description of Implementation Strategy

The European Federation of Neurological Societies has a mailing list and all guideline papers go to national societies, national ministries of health, World Health Organisation, European Union, and a number of other destinations. Corporate support is recruited to buy large numbers of reprints of the guideline papers and permission is given to sponsoring companies to distribute the guideline papers from their commercial channels, provided there is no advertising attached.

# **Implementation Tools**

Staff Training/Competency Material

For information about availability, see the Availability of Companion Documents and Patient Resources fields below.

# Institute of Medicine (IOM) National Healthcare Quality Report Categories

**IOM Care Need** 

Living with Illness

**IOM Domain** 

Effectiveness

Patient-centeredness

# Identifying Information and Availability

# Bibliographic Source(s)

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Not applicable: The guideline was not adapted from another source.

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### Guideline Status

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### Guideline Availability

Electronic copies: Available to registered users from the European Federation of Neurological Societies Web site

### **Availability of Companion Documents**

The following are available:

- Brainin M, Barnes M, Baron JC, Gilhus NE, Hughes R, Selmaj K, Waldemar G; Guideline Standards Subcommittee of the EFNS Scientific Committee. Guidance for the preparation of neurological management guidelines by EFNS scientific task forces revised recommendations 2004. Eur J Neurol. 2004 Sep;11(9):577-81. Electronic copies: Available in Portable Document Format (PDF) from the European Federation of Neurological Societies (EFNS) Web site
- Continuing Medical Education questions are available to registered users from the EFNS Web site

### Patient Resources

None available

### **NGC Status**

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